Inborn Errors of Metabolism

Definition/ cut-off value

Presence of inborn error(s) of metabolism diagnosed by a physician as self reported by applicant/participant/caregiver; or as reported or documented by a physician, or someone working under physician's orders.

Generally refers to gene mutations or gene deletions that alter metabolism in the body, including, but not limited to:

- phenylketonuria (PKU)
- maple syrup urine disease
- galactosemia
- hyperlipoproteinemia
- homocystinuria
- tyrosinemia
- histidinemia
- urea cycle disorders
- glutaric aciduria
- methylmalonic acidemia
- glycogen storage disease
- galactokinase deficiency
- fructoaldolase deficiency
- propionic acidemia
- hypermethioninemia
- medium-chain acyl-CoA dehydrogenase (MCAD)

Participant category and priority level

Category	Priority
Pregnant Women	I
Breastfeeding Women	I
Non-Breastfeeding Women	III
Infants	I
Children	III

Justification

Appropriate dietary management, which may include the use of special formulas, can minimize the medical risk to individuals with inborn errors of metabolism.

Clarifications/ Guidelines

Before assigning this risk code, document the specific inborn error of metabolism on the health history form.

Clarifications/ Guidelines (cont)

The table cited below in reference #3 (Metabolic Disorders Amenable to Nutrition Therapy) includes other conditions not listed in the definition for this risk. If a participant has a physician's diagnosis of a condition not listed in the definition, but included in the table, they may be eligible. If after assessment by a CA, it is determined that the inborn error of metabolism impacts nutritional health and the condition can be ameliorated by WIC participation, the participant can be certified using this risk code. Such case-by-case determinations of nutrition risk do not require Federal approval. However, if a specific condition, not listed in the definition, is frequently used as a certifying nutritional risk, than a request for approval to RISC must be submitted by the State agency.

Self-reporting of a diagnosis by a medical professional should not be confused with self-diagnosis, where a person simply claims to have or to have had a medical condition without any reference to professional diagnosis. A self-reported medical diagnosis ("My doctor says that I have/my son or daughter has…") should prompt the CA to validate the presence of the condition by asking more pointed questions related to that diagnosis.

References

1. Institute of Medicine: WIC Nutrition Risk Criteria: A Scientific Assessment; 1996; pp. 181-183.

2. Queen, PM and Land, CE: Handbook of Pediatric Nutrition; Aspen Publishers. Inc.; 1993; p. 342.

3. The American Dietetic Association: Pediatric Manual of Clinical Dietetics; Table 2-Metabolic Disorders Amenable to Nutrition Therapy; 1998; p. 288.